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Case report

Autoimmune pancreatitis in a patient presenting with obstructive jaundice and pancreatic mass

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Background

Autoimmune pancreatitis (AIP) is a rare cause of chronic pancreatitis.

Case outline

A case of obstructive jaundice with pancreatic mass mimicking malignancy is described.

Discussion

Recognition of the disease by its typical radiological and serological findings permits trial of steroid therapy and may avoid resection.

Keywords

autoimmune pancreatitis, obstructive jaundice, pancreatic mass, steroid therapy

Case report

An abstinent 70-year-old Armenian lady presented with a 2-week history of jaundice, epigastric discomfort, anorexia and 4 kg weight loss. Background history included insulin-dependent diabetes mellitus. Physical examination was unremarkable. Liver function tests were deranged: AST 365 U/L (normal 7–40 U/L), ALT 700 U/L (normal 7–40 U/L), ALP 520 U/L (normal 39–151 U/L), GGT 1640 U/L (normal 6–46 U/L), bilirubin 164 μmol/L (normal 0–18 μmol/L). Abdominal computed tomography (CT) (Figure 1) revealed dilated common and intrahepatic bile ducts, thickened distended gallbladder and a pancreatic head mass.

Endoscopic retrograde cholangiopancreatography (ERCP) showed a 2.5-cm distal common bile duct (CBD) stricture and extrinsic compression of the duodenum. A 10-Fr polyethylene stent was inserted. Endoscopic ultrasound (EUS) confirmed a $3 \times 3 \times 2$ cm irregular pancreatic mass involving the common bile duct (CBD) (Figure 2), with no evidence of portal vein invasion. CBD brushings and EUS-guided fine-needle biopsies were non-diagnostic.

Staging laparoscopy and pancreaticoduodenectomy was performed, and the patient made an uneventful recovery. Histopathology was benign showing severe chronic pancreatitis with sclerosing processes involving the common bile duct and gallbladder, suggesting autoimmune pancreatitis (AIP). Surrounding lymph nodes showed prominent follicular hyperplasia.

Postoperatively, the patient remains asymptomatic with normal liver functions. There is no serological evidence for other autoimmune diseases. Immunoglobulin G (IgG) is 15.5 g/L (normal 6.2–14.4); IgG4 is 0.92 g/L (normal 0.07–0.88). Insulin injections were no longer necessary for diabetic control.

Discussion

Pancreatitis associated with hypergammaglobulinaemia was first reported in 1961, suggesting an immune component to some pancreatic disease [1]. Patients with AIP have raised levels of IgG4 [2]. Hamano *et al.* concluded that AIP was related to the IgG4 immune complex production and that serum levels have a diagnostic role, reflecting disease activity [3, 4]. There may be associations with other autoimmune diseases.

Japan has reported most cases, with males predominant. Mean age at diagnosis is 55 years [5]. Alcohol abuse is uncommon. Clinically AIP may mimic chronic pancreatitis but often with milder symptoms. Obstructive jaundice is common due to narrowing of the CBD within the pancreas [1, 6]. Diabetes mellitus is common (43–68%) [5].

Ultrasound findings include a diffusely enlarged pancreas [1]. Dynamic CT scans display a delayed and prolonged enhancement of the involved pancreatic segments. The pattern may be diffuse or focal reflecting the degree of fibrosis [7]. ERCP changes include irregular pancreatic duct narrowing and CBD strictures. EUS

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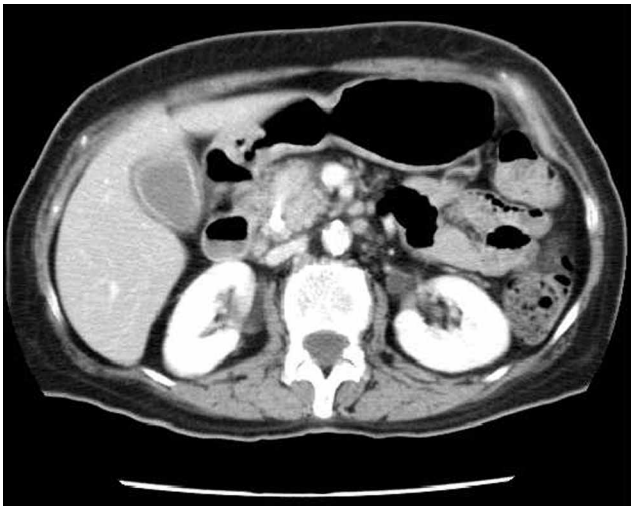

Figure 1. *Pre-operative abdominal CT scan showing a distended and thickened gallbladder and common bile duct, with a biliary stent in situ. The pancreatic head appeared swollen.*

examination of AIP has not previously been recorded, and in our case the complex mass was endosonically indistinguishable from malignancy. Linear EUS permits the possibility of needle biopsy.

Histological changes in AIP differ from alcoholic chronic pancreatitis with predominantly peri-ductal inflammation consisting of a dense interstitial lymphoplasmacytic infiltrate, causing duct obstruction, acinar tissue fibrosis and, occasionally, duct destruction [6, 8]. This process may also affect the CBD, gallbladder and portal vein [8].

Steroid treatment may improve symptoms and pancreatic function with normalisation of secretin tests and blood glucose levels [9], particularly when there is serological evidence of an autoimmune abnormality [7].

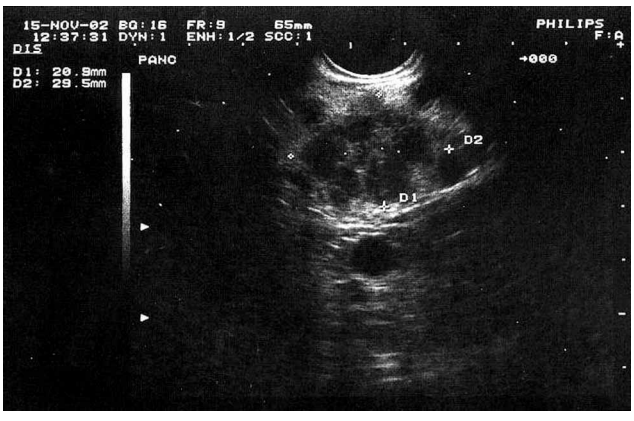

Figure 2. *Endoscopic ultrasound shows a complex mass in head of pancreas.*

Treatment delay may result in irreversible β-cell damage and diabetes despite restoration of exocrine function [4].

Cases are often treated surgically following a provisional diagnosis of pancreatic malignancy. Pancreaticoduodenectomies for benign conditions are relatively common, with an incidence of up to 11% in high volume centres. AIP accounted 27% of these 'benign but clinically suspicious' resections [10]. Preoperative diagnosis of AIP may avoid unnecessary major surgery. However, in patients with a high level of clinical suspicion, but without a preoperative tissue diagnosis of malignancy, resection is justifiable, offering the only hope for cure.

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